Case Report

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Congenital Partial Bronchial Atresia of Right Middle Lobe- A Rare Cause of Dyspnoea

Internal Medicine Section

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ABSTRACT

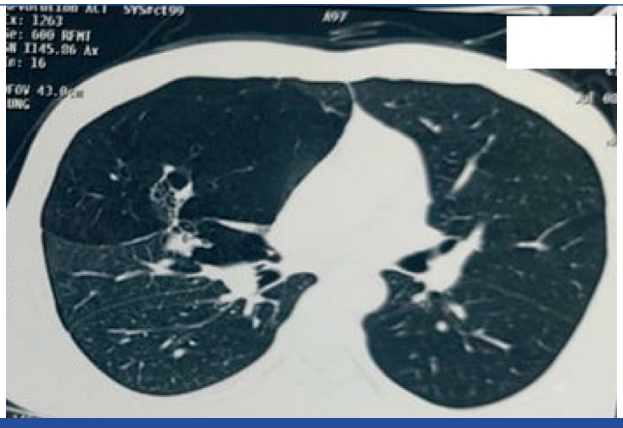
Congenital Bronchial Atresia (CBA) is a rare congenital condition with features of focal obliteration of a proximal segment of a bronchus. Left upper lobe is the most commonly involved lobe. Authors hereby, present a case report of a 17-year-old male who presented with exertional dyspnoea, cough and chest pain. High Resolution Computed Tomography (HRCT) chest showed hyperventilated lung parenchyma with paucity of bronchovascular pattern in right middle lobe. Bronchoscopy revealed right middle lobe bronchial segmental atresia with rudimentary bronchus above that segment. The patient was treated with bronchodilators, steroids and improved symptomatically and advised surgical management. Patient was not willing for the same and hence managed conservatively. Though definite management for CBA is done surgically, medical management can be preferred for patients with mild symptoms and without recurrent infections.

Keywords: Bronchoscopy, Computed tomography chest, Rhonchi

CASE REPORT

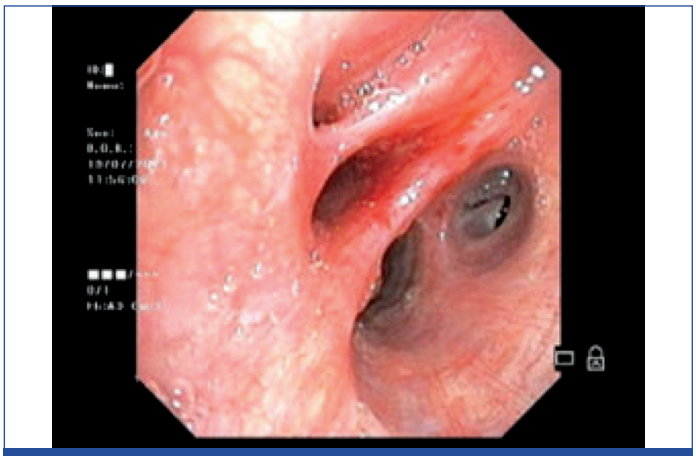
A 17-year-old male with no co-morbidities, complained of shortness of breath on exertion, dry cough and increased right-sided chest pain for 15 days and one episode of low grade fever 2 days back. He had similar complaints for the past 3 years with varying severity and was managed in different hospitals with various antibiotics (amoxicillin with clavunic acid, levofloxacin, and cefpodoxime), steroids (prednisolone), bronchodilators (salbutamol) during previous episodes and not on regular treatment at the time of presentation to the hospital.

The patient had no significant family history/seasonal variation of symptoms. His vitals were stable and were afebrile during presentation. Auscultation revealed decreased breath sounds in right mammary region with occasional rhonchi. Routine investigations, electrocardiogram and echocardiography were found to be normal. Chest radiograph showed hyperlucency in the right mid and lower zone. High resolution Computed Tomography (HRCT) chest showed hyperventilated lung parenchyma with paucity of bronchovascular pattern and mucocele in right middle lobe suggestive of bronchial atresia [Table/Fig-1]. Spirometry was found to be normal.



[Table/Fig-1]: Hyperventilated lung parenchyma with paucity of bronchovascular pattern in right middle lobe with impacted mucocele.

Bronchoscopy showed right middle lobe bronchial segment partial atresia with a rudimentary bronchus above it, bronchoscope could not be passed through the segment and all other segments were normal with no endobronchial lesion visualised [Table/Fig-2]. Bronchoalveolar lavage did not show any growth in the culture. The above presentation of symptoms and investigations confirmed the diagnosis of right middle lobe segmental bronchial atresia. The patient was started empirically on nebulised salbutamol 2.5 mg every 6th hourly, nebulised budesonide 0.5 mg every 12th hourly injection amoxicillin 1000 mg+clavulanic acid 200 mg twice a day for 5 days and injection methylprednisolone 40 mg once a day was required for five days in view of persistent rhonchi inspite of adequate nebulisations. Patient improved symptomatically after five days of therapy and was advised surgical management. Patient was not willing for the same and was on regular follow-up. He had no new complaints and was symptomatically better in his last follow-up on September 2021.



[Table/Fig-2]: Bronchoscopy showing right middle lobe partial atresia with a rudimentary bronchus.

DISCUSSION

The Congenital Bronchial Atresia (CBA) is a rare congenital condition which was first reported by Ramsey BH, in 1953 as focal interruption of a lobar, segmental, or subsegmental bronchus with hyperinflation and mucus impaction of the obstructed lung segment [1,2]. There are two types of CBA- proximal type and peripheral type. The proximal type affects the lobar bronchus. It is extremely rare and usually fatal during pregnancy and causes a massive increase in the volume of the distal lung with secondary hypoplasia of the normal parenchyma. The peripheral type affects the segmental or subsegmental bronchus as similar to this patient [3].

Bronchial atresia is usually diagnosed in the twenties or thirties with predominance of males, with a prevalence of 1.2 cases per lac males as per Psathakis K et al., [4]. It is more common in males and the mean age of diagnosing CBA is 34.7 as per Puglia EB et al., [5].

The exact cause of CBA is not known. The pathogenic mechanism of CBA that is widely accepted is an "accident" to the nutrient artery of the bronchus that leads to ischaemia and atresia of the bronchus [6]. The left upper lobe is most often involved, followed by other lobes [4,5].

Most of the bronchial atresia patients are asymptomatic and only a few will have symptoms such as cough, dyspnoea and haemoptysis [4]. Decreased breath sounds on auscultation of the affected area is the most frequent clinical finding similar to this case, there was diminished breath sounds over right mammary area [7]. Symptomatic patients usually present with recurrent pulmonary infections, wheeze and dyspnoea similar to this patient.

Bronchial atresia is an incidental finding on chest radiograph and Computed Tomography (CT) is the gold-standard method for establishing the diagnosis of CBA [8]. The main CT findings include area of hyperinflation distal to the affected bronchial segment, air trapping and mucocele which was exactly seen in this patient [8]. In few patients, Magnetic Resonance Imaging (MRI) is also used to see for arteriovenous malformations [4]. As in this case CT images showed mucocele surrounded by a hyperlucent lung representing air trapping which is believed to occur by collateral ventilation through alveolar pores of Kohn, interbronchiolar Martin's channels and bronchoalveolar Lambert's channels as per Terry PB and Traystman RJ [9].

A typical mucocele is usually found distal to the point of atresia in the absence of infection, as in this case, the adjacent parenchyma can be normal or hyperinflated [10]. Microscopic analysis shows distended alveoli with minimal or absent destructive changes which is the major difference between emphysema and related disorders (congenital lobar emphysema) in which there will be destruction and expansion of alveoli [10].

The differential diagnosis for mucus impaction include cystic fibrosis, allergic bronchopulmonary aspergillosis, bronchial cyst, pulmonary embolism, bronchiectasis, or intralobar sequestration any bronchial narrowing causing mucus impaction, but the presence of a mucocele along with hyperinflation helps to narrow down the diagnosis. In adults bronchial carcinoids should also be considered as a differential diagnosis of bronchial atresia, as bronchial carcinoids may also cause air trapping and mucoid impaction for which contrast enhanced CT or MRI helps distinguish bronchial atresia from carcinoids as the latter may show central enhancement within the mucoid impaction. Moreover, the signal intensity of carcinoids may not be as high as that of mucoceles on T2-weighted MRI [11].

Bronchoscopy is usually done when classical CT features are present and to rule out alternative diagnosis like endobronchial obstruction, foreign body or tumour as in this case endobronchial obstruction ruled out but the bronchoscope could not be passed through the atretic segment like in this case [2,12].

Recurrent infections and pneumothorax are the major complications of CBA and rarely pulmonary hypertension is seen as a long term sequel due to CBA [13]. CBA complicated by a lung abscess due to *Aspergillus fumigatus* has also been reported and treated with voriconazole and resection [14].

Treatment of CBA is can be conservative or surgical. Surgery is indicated in patients with recurrent and severe respiratory symptoms like haemoptysis, dyspnoea, cough and pneumonia and if medical treatment is ineffective. The surgeries usually done are lobar resection and segmentectomy [15]. The ultimate aim of surgery is to preserve maximum normal lung parenchyma to maintain pulmonary function for the patients [5].

CONCLUSION(S)

Congenital bronchial atresia though a rare entity should be suspected in a young age group especially males with obstructive airway symptoms and diminished breath sounds in a localised area during examination. Computed tomography is the first modality of choice to diagnose it. If patients are asymptomatic, conservative management can be the first approach. Surgical intervention is required for definite management and when there are recurrent infections or compromise of adjacent lung parenchyma. CBA can also present with complications like pneumothorax, pulmonary hypertension, *aspergillus fumigatus* and should be treated accordingly. Early resection should be offered for all the adults diagnosed with CBA for definite management keeping in mind the long term possible complications.

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